HEREDITARY SPASTIC PARAPLEGIA.[1]

By WESTON D. BAYLEY, M.D.

IN the family about to be described, it is known that five generations have presented cases of primary lateral sclerosis. How much more remotely this singular malady extended it is impossible to learn.

The genealogical tree here shown represents the one accessible branch of the family as far back as can be authentically traced. Other members of the fourth generation and their descendants are known to have been afflicted in like manner to the one here described, but no authentic information from them is at this writing obtainable. For assistance in the collection of these records, I am indebted to my friend, Dr. F. Mortimer Lawrence, who with me made personal examination of the appended cases.

The numericals on the diagram indicate the cases personally examined, and identify them in the records which follow.

H. F., unmarried, female, æt. 31, housewife (Case 1 of tree). Apparently well until five years of age, when she gradually became paraplegic, showing at first a spasticity of the adductors of the thigh. The trouble has been symmetrical from the start. At no time has there been pain, numbness or other sensory symptoms; no bowel or bladder difficulty, no atrophy, no static ataxia, no diplopia, no deformity of the feet. Muscles hold the limbs rigid. Knee-jerks are apparently absent, even with re-inforcement. Pupils equal—both react to light, but the right a little more promptly than the left.

C. F., married, male, æt. 28, telegraph operator (Case 2 of tree). Apparently well until five years of age,

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[1] Read before the A. R. Thomas Club, of Philadelphia.

- Female afflicted.
  - Male afflicted.
    - Male unafflicted.
      - Male unafflicted.
      - " "
      - Female "
      - Male "
      - Female "
      - Male "
    - Female unafflicted.
      - Female unafflicted.
        - Female unafflicted.
        - Female unafflicted.
      - " "
      - " "
      - " "
    - Female afflicted (unmarried).
    - Male afflicted (died æt. 55 of apoplexy).
      - Female afflicted. 1.
      - Male " 2.
        - Male afflicted. 9.
        - Male unafflicted æt. 2 years.
      - Female (died in convulsions æt. 3 weeks.
      - Male unafflicted.
        - Male unafflicted.
      - " "
      - Female afflicted. 3.
      - Male " . 4.
      - Female unafflicted
      - Male afflicted. 5.
    - Male unafflicted.
      - Male unafflicted (died in infancy of hip joint disease).
      - Male unafflicted.
      - Female "
      - Male "
      - Male "
      - Female "
      - Male "
      - Female "
    - Female (died in infancy).
    - Female unafflicted.
      - Female (neurotic) unafflicted.
        - Female under 3 yrs. unafflicted.
      - Female unafflicted.
    - Male afflicted. 6.
      - Female unafflicted (neurotic, choreic, etc.).
        - Female under 3 yrs. unafflicted.
      - Female (died æt. 9 months of sp. meningitis.
      - Male unafflicted.
      - " "
      - Female afflicted. 7.
      - Male " . 8.
    - Male unafflicted.
      - Female unafflicted.
        - Female unafflicted.
        - Female unafflicted.
      - Male "
      - Male "
      - Female (died 1 yr. of measles.
      - Female unafflicted.
      - Male "
      - Male "
      - Male "
  - At least one afflicted brother of whom no definite information is at present obtainable. Descendants of him or his brothers or sisters are known to be afflicted.

when his "legs began to give out." The first symptom appeared to be hyper-adduction of the thighs, and later general spasticity of both lower extremities. This condition has persisted without material change to the present time. Paretic spastic gait with contractures of the tendo-achillis. Knee-jerks are exaggerated, pupillary reflexes normal and very free, ankle-clonus on both sides. Has never had pain or numbness; no static ataxia; bowel, bladder and sexual reflexes are normal.

E. F., unmarried, female, æt. 19, housewife (Case 3 of tree). Apparently well until between her fifth and sixth year, when adductor spasm, soon followed by spastic paraplegia, developed. No pain or numbness. Knee-jerks very excessive, the slightest touch producing a vigorous response. Well-marked ankle-clonus in the left foot only. No diplopia; pupils react normally; no bowel or bladder symptoms; no static ataxia.

T. F., unmarried, male, æt. 16, schoolboy (Case 4 of tree). No illness until the fifth year, when spasticity of the legs developed, with adductor spasm without sensory symptoms. Knee-jerks excessive, marked ankle-clonus, no static ataxia, talipes equinus. Pupils equal and react normally. No bladder or bowel symptoms.

C. F., male, æt. 11, schoolboy (Case 5 of tree). No symptoms until his third year, when the spastic gait appeared in the same manner as with the others. Talipes equino-varus; knee-jerks free, slight ankle-clonus in both feet; the deformity has been progressive during the past year. Pupils equal, and react normally, no static ataxia, no bowel or bladder symptoms, no sensory symptoms.

C. F., married, male, æt. 51 (Case 6 of tree). Was apparently well until between his fourth and fifth year, when spasticity of gait appeared, being most marked at first in the adductors of the thigh. There developed early tendo-achillis contracture, and because of the deformity, tenotomy was performed twenty-three years ago. Since this operation he has not been able to walk nearly so well as before it. The knee-jerks are very excessive; no ankle-clonus because of the tenotomy. No sensory symptoms; no bowel or bladder difficulty. Pupils react normally; no static ataxia.

The two following cases are children of Case 6.

M. F., female, æt. 13, schoolgirl, (Case 7 of tree). Apparently well until her seventh year, when her previously normal gait became disordered by adductor spasm, and the spastic gait developed. No sensory symptoms at any time. Knee-jerks excessive; marked ankle-clonus on both sides; no static ataxia. Pupillary reflexes normal; no bowel or bladder difficulty; tendency to talipes equinus.

R. F., male, æt. 7 (Case 8 of tree). Was well, and, as was the case with the others, walked normally until his sixth year, when the same character of spastic gait developed, with the early symptoms of adduction, causing "cross-legged progression." Knee-jerks excessive; slight ankle-clonus. Pupillary, bowel and bladder reflexes normal; no static ataxia. As with the other cases, never any complaint of pain, numbness or anæsthesia.

H. B. F., male, æt. 2 years (son of Case 2, Case 9 of tree). "Never learned to walk right." Has had the spastic tendencies from the earliest efforts to walk, but less marked than at present. He falls easily, and walks with a "spastic waddle." Has difficulty in arising from the floor, but does not "climb up on himself." No muscular atrophy. Child is well nourished; bright mentally; knee-jerks are excessive; no ankle-clonus.

The group of symptoms presented by these cases clearly places the lesion as a primary sclerosis of the crossed pyramidal tracts. The wonderful similarity of the cases, both as to time and mode of onset, absence of sensory phenomena, and (excepting in the first case) state of reflexes, established its hereditary type. It is obvious, therefore, that another congenital affection must be placed in the lists with the already well-known Friedreich's ataxia and pseudo-hypertrophic paralysis. From these two hereditary diseases it differs in the most obvious manner. Nor has it more than a cursory resemblance to the spastic paraplegia of infancy, which is cerebral in origin, involves the upper extremities with the lower, presents cerebral symptoms, and is believed to be absolutely non-hereditary.

It would be of great interest to know the pathological condition in these cases; whether the lesion originates in a vascular defect or is primarily inherent in nerve tissue. This, in the absence of opportunity to examine the cord, must remain conjectural. It is worthy of note, however, that in this family the escape of an individual from the affliction seems to confer immunity upon his or her descendants, the affection travelling in direct and not in collateral lines.

HEMIATROPHIA TOTALIS CRUCIATA. (Deutsche medicinische Wochenschrift, No. 12, 1897.) By M. A. Lunz.

The peculiar affection reported in this paper, dated from the patient's marriage and exposure in field work. It began with periodic pain in the posterior part of the right thigh, which radiated downward into the sole of the foot. After three years, or six years before the report was made, the patient observed wasting of the right lower limb; she had also noticed that the right breast was smaller than the left, and secreted less milk. Three or four years ago the left cheek was observed to be smaller than the right, and at this time the patient began to complain of a pain in the cheek. For a year she had had pain in the back and left lower limb, as well as in the region of the right scapula and right upper limb. At the time of examination by Lunz, the wasting in the skin and muscles of the left cheek was very perceptible. The circumference of the shoulder at the axilla was 43 cm. on the left side, and 41.2 cm. on the right, whereas at the insertion of the deltoid there was no difference on the two sides. On the posterior part of the thorax, atrophy was noticed only at the inner border of the scapulæ. The atrophy on the right side of the abdomen was very perceptible, but still more so in the right half of the pelvis and in the right lower extremity. All movements were well performed, and there was no trace of paresis, but the right leg became tired sooner than the left in walking. The pain was increased by movement. Pressure over the lower lumbar and sacral vertebræ, and over the nerves and muscles of the right lower extremity, caused, at times, sensation of pain. Objective sensation otherwise was not altered. There was no reaction of degeneration. The patellar reflexes, especially the right, were somewhat exaggerated. The process could not have been central, as the hyperesthesia and atrophy were limited to certain regions innervated by certain nerves. It was evidently a neuritis migrans.

Irritation of sensory nerves acts on the vasomotor centres in such a manner that vascular constriction is produced. Attacks of headache, vertigo, tinnitus aurium, sensation of heat, free perspiration, as well as subjective sensation of cold, and the livid appearance of the lower limbs, observed in this case, were probably due to vasomotor disturbance. The writer believes that the results obtained in noting the surface temperature of the affected and sound parts of the body are in favor of the theory of vasomotor disturbance. SPILLER.